ordinarily associated with endocarditis, and where there is evidence of well-marked arterio-sclerosis.

A Flint murmur may, however, be associated with many of the clinical features of a true organic mitral obstruction.

OSTEITIS DEFORMANS.[1]

By Frederick A. Packard, M.D., J. Dutton Steele, M.D.,
and
Thomas S. Kirkbride, Jr., M.D.,
of Philadelphia.

Before proceeding with the report of this case we would state that the histological examination of the tissues was begun by the late Dr. T. S. Kirkbride, Jr., whose untimely death from typhoid fever, contracted in laboratory work, came soon after the beginning of what promised to be a brilliant career. The work so begun by Dr. Kirkbride was taken up and carried to completion by Dr. Steele.

In April, 1899, there was admitted to the men's ward of the Philadelphia Hospital a German, aged sixty-two years, who for some months had been an inmate of the almshouse department. Until the onset of an acute indisposition, for which he was referred to the wards, he had attracted no particular attention. Owing to the fact that he could speak almost no English and was very deaf, with some impairment of mental power, nothing could be learned of his past history. Attention was attracted to him by the size of his head, which was that of a man very much larger than he was. Not only was his head large, but it had a peculiar shape, being rather box-shaped, with a marked projection of the forehead, flattened crown, and a distinct tumor at the junction of the left temporal, frontal, and parietal regions. (See Figs. 1, 2, and 3.) The face was somewhat larger than that of a normal individual of the same size, but was small in proportion to the size of the calvarium. The mastoid processes were large and heavy, and the lower jaw was also disproportionately large, but was not prognathous. The soft parts of the face showed nothing out of the way. Measurements of the head were taken shortly after his admission, and again about a year later. For the purpose of comparison both sets of measurements are inserted here:

	Shortly after admission.	February 12, 1900.
Bitemporal	14 cm.	15½ cm.
Binaural	15 "	14½ "
Fronto-occipital	20 "	20¾ "
Occipitomental	20 "	23 "
Biparietal	17 "	19½ "
Angular	13 "	13 "
Greatest circumference . .	63 "	63 "
Circumference of vault from meatus to meatus . . .	40½ "	41 "

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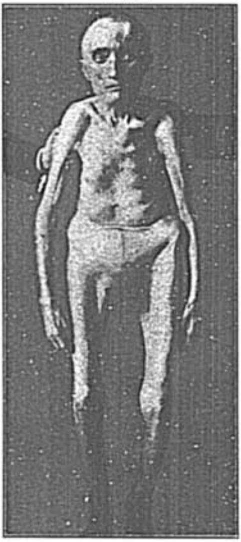
[1] Read before the Association of American Physicians, 1901.

The head and face, therefore, were rather egg-shaped, with the larger end of the oval pointing upward, as is well shown in the figures. The tongue was of normal size, the vault of the hard palate capacious, but not deep; the alveolar processes were excessively thick. All of the sutures and fontanelles were closed, and showed no depression or ridging. The speech was very peculiar, and practically unintelligible, although not entirely because of ignorance of the language, as an occasional English word could be discerned so thickly emitted that its recognition was almost impossible. The larynx was very large and broad, giving the neck a rather full appearance, but the thyroid gland could not be outlined.

A curious alteration of the shoulder girdle was present, due to the fact that, while the clavicles stood out prominently because of thickening of the bones and increase in their natural curves, their direction was almost backward. The upper ends of both humeri, especially the heads, were much thickened, and the natural curves and elevations were very much exaggerated. On both sides there was marked anterior curvature at the level of the deltoid insertion. The elbow-joints appeared to be normal, as were also the bones of the left forearm. In the right forearm there was marked curvature of both bones toward the radial side. On both sides the radius and ulna had a distinctly rough and uneven surface. The appearance of the chest can best be appreciated by a glance at Fig. 2. Above, the chest was small and narrow with the antero-posterior diameter almost equal to the lateral, while below the chest was large and bulging. No substernal dulness could be detected. The xiphoid process was very prominent and broad, and the ribs were distinctly thick and heavy. On each side the tip of the eleventh rib was almost in contact with the crest of the ilium. The costo-sternal angle was very obtuse. There was marked forward curvature of the spine, with very evident thickening of the lower dorsal and upper lumbar vertebræ. Dorsocervical kyphosis was present without scoliosis. The scapulæ were apparently not enlarged.

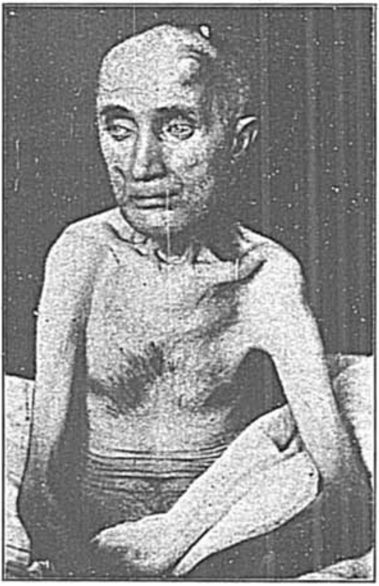
FIG. 1.



Photograph of subject of report, showing shape of head, frontal tumor, and bony deformities.

Examination of the abdomen showed that there was excessive pigmentation of the skin and growth of hair on the anterior surface. Just beneath the costal margin there was a deep transverse furrow extending across the abdomen. The general outline of the abdomen was that of a diamond, the angles of which were formed by the xiphoid angle, the pubis and the junction of the ribs with the crests of the ilium. The pelvis resembled in shape that of the female rather than that of the male, being very broad and heavy, and the superior basin being very capacious. The distance between the anterior superior spines was 31 cm. The iliac crests were remarkably heavy, and projected far beyond the level of the trochanters. The extreme lateral width of the pelvis was 34 cm., the intertrochanteric distance being 30 cm., while the patient's total length was only 57 cm. The thigh bones were somewhat large, but were not much deformed, except for the condyles, which were very large. The patellæ were about normal in size. In the skin over the left patellar tendon there was a flat, circular mass of about 2.5 cm. in diameter, apparently situated in the superficial layers of the skin, and giving to the finger the sensation of an enchondroma. Both

FIG. 2.



Photograph of subject of report, showing size of head, frontal tumor, clavicle, and peculiar shape of chest.

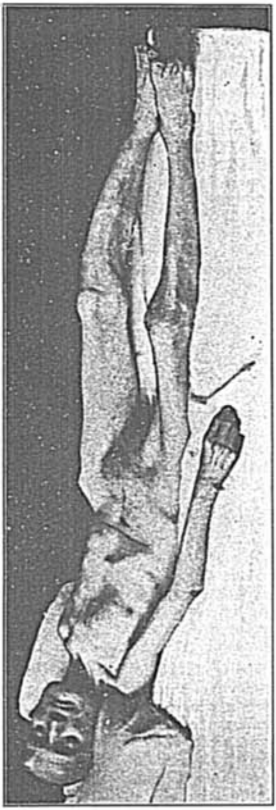
tibiæ were greatly increased in thickness, especially on the anterior aspect, the surface of which was rough. This thickening and prominence were more marked on the left side than on the right. The fibula of the left side was decidedly thickened. Both legs showed marked outward bowing, the distance between the knees when the feet were placed together being 7.5 cm. The hands and feet were not enlarged.

In no part of the body was there evidence of involvement of the soft parts. The whole attitude of the patient on standing up irresistibly reminded one of that of the orang-outang.

On his admission the man had a large pleural effusion in the right side. This was withdrawn, and did not reaccumulate. He, however, remained feeble with progressively increasing emaciation and slow growth of the tumor in the frontal bone until his death, about a year after coming under observation.

Autopsy was held twenty-four hours after death. The following notes were made at the time: Rigor mortis is well marked. External conformation as described in clinical notes. On removing the scalp there is found a large subcutaneous tumor springing from the frontal bone to the left of the median line. The tumor is of soft consistence. In places the scalp is remarkably thin, especially in the left temporal region. The outer surface of the calvarium has a rather bluish appearance, and seems somewhat more vascular than normal. The saw cuts very easily through the bones of the skull. The tumor mentioned above is found to measure 4 x 5 cm. at its base, and has an elevation of 2 cm. The calvarium is fairly uniformly increased in thickness, measuring from

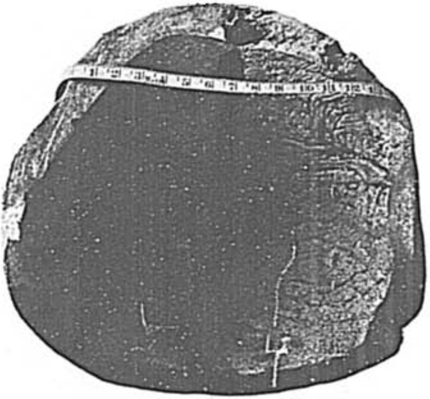
FIG. 8.



Showing especially the separation of the knees when the ankles were in contact.

1½ to 2 cm. (See Fig. 4.) The diploë is apparently absent, excepting in the frontal and occipital regions. A little to the left of the median line, at a point corresponding to the superficial tumor, the bone is thickened to 3 cm., owing to the presence in the diploë of a soft growth corresponding in consistence to that seen externally. The growth projects through both the inner and outer tables. The brain is adherent to and almost incorporated with the growth. The whole hollow of the frontal bone on the left side is occupied by a large irregularly rounded mass, of soft consistence, and measuring 5 cm. in all directions. The base of the skull appears large and gross, and all of its concavities are deepened except the sella turcica, which is of about normal size. The inner table of the calvarium is deeply grooved for bloodvessels and freely perforated by small foramina. The base of the brain shows great overfulness of the bloodvessels. The membranes of the brain seem unaltered over a greater part of the surface, but over the convexity the pia is slightly opaque. In the left frontal region anteriorly to the operculum the brain substance is torn and softened at a point corresponding to the tumor of the calvarium. On the posterior end of the third frontal convolution there are some small whitish granules (sarcoma or tuberculosis ?). Similar masses are found bordering upon the longitudinal sinus of the left side.

FIG. 4.



Calvarium.

On the right side there are some old adhesions of the pleura, especially near the apex ; the left pleura contains a small amount of fluid. The layers of the pleura on the left side are much thickened, and there is on them a gelatinous substance which partially fills the cavity. A large " milk patch " is present on the anterior wall of the left ventricle. The heart muscle is soft and flabby, but there is no other alteration of the walls or valves. There is an area of atheroma at the root of the aorta. The left lung is almost but not quite airless. The diaphragm on the left side shows irregular thickenings of yellow color looking like sarcoma. The right lung shows only hypostatic congestion. The spleen is large, measuring 13 x 8 x 3.5 cm. The pulp is very soft and the Malpighian bodies prominent. The left suprarenal capsule is thin, and the distinction between the cortex and the medulla is very poorly marked. The kidneys are large and pale, with somewhat prominent vessels, and capsules stripping off readily. The right suprarenal resembles the left except for the fact that there is a postmortem cavity in the medulla. The liver shows slight distention of the small bile passages; the substance of the organ appears fairly normal. The bile ducts are patulous, and the mucous membrane of the gall-bladder has a normal appearance. The pancreas is rather small, but seems otherwise normal.

Owing to the limited time allowed for the autopsy, it was impossible to remove the spinal cord or the peripheral nerves. It is much to be regretted that preservation of the thyroid gland was for some reason inadvertently omitted. Grossly it showed no change, but it would have been advantageous to have proved its condition by microscopical examination. While changes in the spinal cord described by some observers are probably, as they have pointed out, due to the accompanying arterio-sclerosis, our inability to remove the spinal cord because of lack of time is a source of regret.

Microscopical Examination.—Clavicle. A portion of the bone was taken from about the middle of the shaft of the thickened portion. It was cut with the saw with somewhat greater ease than a normal bone. Decalcification in 5 per cent. nitric acid and phloroglucin was complete in twelve hours, showing the small amount of lime salts in the tissue, as ordinary bone requires from three to four days for decalcification in this solution.

The sections were stained by hæmatoxylin and eosin. The part of the shaft that is usually compact bone substance, with regular Haversian systems, consists of a finely porous structure in which areas of unaffected bone alternate with spaces that are evidently the result of its absorption. These spaces are filled with a fibrous-tissue reticulum. The cells of the spaces are mostly spindle in character; but there are also many round cells, with deeply staining nuclei, polymorphonuclear leucocytes, and a few nucleated red cells, and some fat cells. Most of the interspaces are exceedingly vascular. In many there are networks of dilated capillaries whose walls are but a single layer of endothelium. Larger arteries cannot be seen in the sections obtained. (See Fig. 5.)

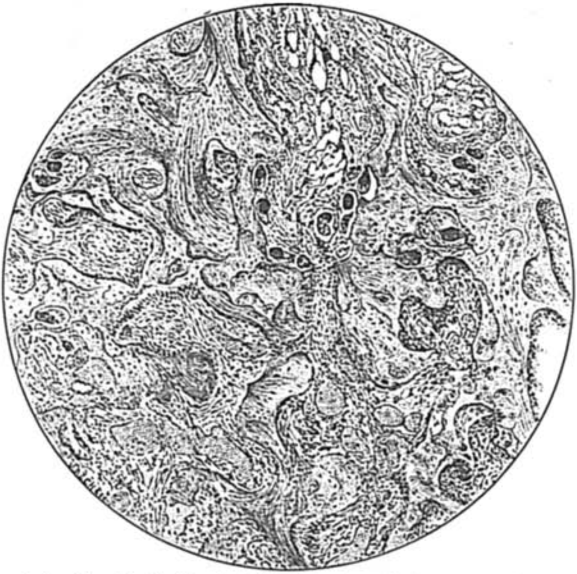
The reticulated structure runs almost to the periosteum, and a thin shell immediately under it is the only representative of the compact substance.

In the interspaces are numerous groups of giant cells with many nuclei. These cells lie in the edges of the unaffected bone, and are apparently engaged in its absorption.

Running through these interspaces and through the bone itself are masses of closely packed cells, resembling osteoblasts, with a fibrous matrix. This is evidently an attempt at the formation of new bone, but in no place is it calcified. The dividing line between the unabsorbed and new bone is sharp, and there is every indication that the processes are distinct from one another.

Skull. The distinction between the diploë and outer and inner tables is entirely lost, as far as could be seen from the sections of the bone obtained. There is a thin and irregular plate of compact bone under the periosteum, but elsewhere the bone is honeycombed by absorption spaces, giving it a porous structure. These interspaces (as in the clavicle) are filled by a connective-tissue reticulum containing spindle cells, round cells with large nuclei, polynuclear leucocytes, and a few giant cells. This last variety are not as numerous as in the sections from the clavicle. As in the clavicle, there are bundles of osteoblasts in a fibrous matrix running through the bone and spaces in all directions, evidently representing new bone. The interspaces in the skull are not as vascular as in the clavicle; however, they are well supplied with blood by capillaries. No large arteries are seen in the section. Sec-

FIG. 5.



Section of the shaft of the diseased clavicle. Leitz ocular 3, objective 3. The general asymmetry, absorption spaces and the fibrous, medullary substance, containing giant cells and many bloodvessels, are well shown. There are several areas of uncalcified new bone.

tions from the tumor of the frontal bone show it to be a typical giant-celled sarcoma. The mass of the tissue consists of bundles of spindle cells and masses of round cells, but there are also many larger cells with a single, irregularly staining nucleus, and numerous giant cells with five or more nuclei. In general, the sections examined show the following changes:

1. Absorption of the healthy bone.
2. Formation of new bone coincident, but in no way connected, with the absorption process.
3. The failure of calcification in this new bone.
4. The destruction of the regular structure of the bone and the addition of new uncalcified bony tissue.
5. The formation of a giant-celled sarcoma in the affected portion of the skull.

These changes correspond closely to those described by Paget, Stilling, and others who have subjected their cases of osteitis deformans to a microscopic examination.

Suprarenal Bodies. Microscopic examination shows these structures to be normal.

Hypophysis Cerebri. Sections of the hypophysis cerebri show no abnormality.

Nodule from the Pia. Consists of a collection of round and spindle cells with a moderate amount of intercellular substance and with deeply staining nuclei. It is well supplied by bloodvessels whose walls consist merely of a single layer of endothelium. In many places giant cells are found. The nodule is evidently a metastasis from the tumor of the frontal lobe.

The nodule from the diaphragmatic pleura is of the same structure as the nodule in the pia, and has evidently the same origin. It also shows numerous giant cells.

In the literature we find references to ninety-nine cases reported as osteitis deformans.[1] Careful study of these cases shows, however, that four of them were certainly instances of some condition other than even slightly developed osteitis deformans. Twenty-nine may possibly have been instances of this disease, but the facts given were not sufficiently definite to warrant the statement that they were typical cases of osteitis deformans. Many of these reported as instances of this disease were apparently examples of tumors of bone of more ordinary nature, or of osteomalacia or fragilitas ossium, with local enlargement secondary to fractures. Sixty-six cases were found which, in our judgment, are typical instances of the disease described by Paget, in 1877, in an article in the *Medico-Chirurgical Transactions.*

ETIOLOGY. Among the cases so far reported, 41 (including our own) were males, 24 females, and in 2 the sex is not stated. The youngest patient at the time of coming under observation was that of Watson, whose age was thirty-nine years, while 2 patients (Silcock's and Lloyd's) were eighty-two years old when they were first seen. Moizard and Bourges have, however, reported a patient who was seventy-three years of age when coming under observation, but the disease seems to have quite definitely started at the age of twenty-one. The average age at the time of coming under observation was sixty-one years; the age at the time of onset was capable of determination in 51 cases, and averaged forty-nine and one-half years, the youngest of these being the patient just mentioned, the oldest patient at the time of onset being between seventy-nine and eighty years of age. As will be seen, there is a marked difference between the time of coming under observation and the time when the first symptoms of the disease were noted; 11 of the 51 cases noted had symptoms definitely pointing to the onset of their disease when they were forty years old or younger. It can, therefore, hardly be said to be exclusively a disease of those very advanced in years.

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[1] Since the presentation of this paper a case has been reported by Atkinson (Maryland Medical Journal, July, 1901). The patient was a male, aged thirty-seven years. The bones affected were the skull, left forearm, and both femora, with considerable deformity. There was no autopsy. The case is apparently one of true osteitis deformans.

The mental capacity of our patient was but slight. In 1 case from the literature (Joncheray's) the patient and one other member of the family suffered from insanity, while a patient of Lunn's was insane. Inheritance seems to play but a small part in the etiology of the disease, although among the cases which we have studied we found two brothers with the disease reported by Lunn, while Pic and Robinson have each reported a case where the disease was present in two members of the same family.

The etiology of osteitis deformans is, therefore, very uncertain. It is undoubtedly a disease of advanced life, although, as we have mentioned, in eleven of the cases collected the disease began before the age of forty, while in the case of Moizard and Bourges the disease began at as early an age as twenty-one years. The influence of heredity is doubtful. In reviewing the literature there seems no striking evidence of any marked influence exerted by consanguineous marriages, nor does there seem to be any marked neurotic tendency in the families of those suffering from this disease. While a large proportion of the cases are found in English journals, too much importance should not be attached to locality as an etiological factor, as the disease was first described by Paget, and we would expect his countrymen to be more familiar with the disease described by him than those whose attention had not been particularly drawn to the subject. In this connection it is interesting to note how frequently a series of cases are reported in rapid succession in various countries, showing that the mere fact of the public mention of the disease would seem to excite interest in the condition sufficient to cause recognition of other cases.

SYMPTOMATOLOGY. The analysis of the cases reported shows that in 14 enlargement of the head was first noted; that in 13 deformity of both tibiæ was mentioned as the first sign of the disease; in 11 the left and in 6 the right tibia was first involved; while deformity of the spine was first seen in 2; of the right radius in 2; of the right clavicle, lower jaw, and sternum each in 1 case.

Analysis of the cases in regard to the bones affected at the time of coming under observation was not possible in all of the cases. In those where sufficient details were given it was found that deformity was distributed as follows: In 49 of the cases the bones of the skull were involved. (It should be noted that among the 66 cases from the literature 9 departed from the type described by Paget, in the fact that there was no manifest enlargement of the head, 1 of these cases having been seen by Paget and considered by him a true instance of the disease.) Both tibiæ were affected in 47, the right tibia alone in 2 cases, the left tibia alone in 1 case. In 40 cases both femurs were affected, while the right femur was alone involved in 3, the left femur in 1 case. The spine was affected in 31 cases when they came under observation. In 24 cases both clavicles were thickened and deformed, the right clavicle being affected alone in 3 cases, the left in 4 cases. The bones of the pelvis were affected in 21 cases. In marked contrast to the frequency of involvement of the tibiæ, the fibulæ were found to be affected in only 10 cases. The humeri were symmetrically involved in 14 cases, while

FIG. 6.

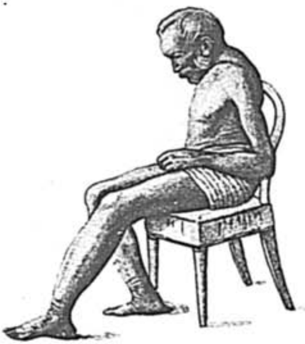

FIG. 7.

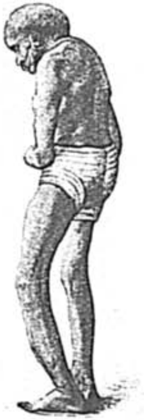

Fig. 6.—From Paget's original paper. (Medico-Chirurgical Transactions, 1877.)
Fig. 7.—From Paget's original paper. Ibid.

in 3 cases that of the right side and in 4 that of the left side were involved. Both radii were affected in 11 cases, while in 5 the right alone and in 3 the left radius alone were affected. In 10 cases the ulnæ were involved, the left ulna being deformed in 2, while the right ulna was noted in no case as being affected where the left escaped. The scapulæ were involved in 6 cases. The ribs were thickened in 16, while the sternum was involved in 7. In 8 cases it is mentioned that the lower maxilla was apparently affected by the disease, while in 1 case it is stated that the bones of the face were involved. In only 8 instances were the patellæ affected. The metatarsal bones were involved in 5, the tarsal bones in 2 cases, and the calcaneus in 1 case. It is seen, therefore, that after the skull the tibiæ are the bones most frequently affected, these being followed by the femurs, spine, pelvis, and clavicles, ribs, radii, and ulnæ. This distribution would seem to rather point

FIG. 8.

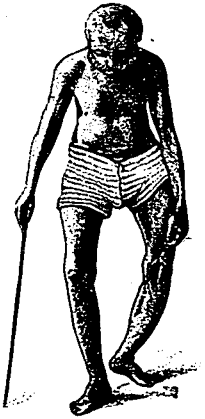

FIG. 9.

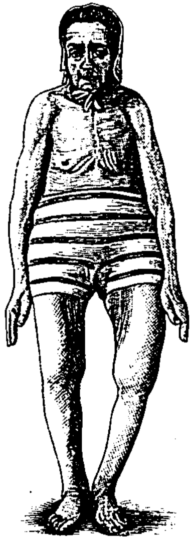

Fig. 8.—From Paget's original paper. (Medico-Chirurgical Transactions, 1877.)
Fig. 9.—Marie's case. (Nouvelle Iconographie de la Salpêtrière, 1894.)

toward the fact that pressure and gravity had at least a large share in the production of the deformities.

While in many of the cases there has been complaint of pain in the bones subsequently affected, our study of the literature would not cause us to attach much importance to this as a symptom of the disease, nor, in fact, has the latter much symptomatology, the results being almost entirely objective.

Fig. 10.

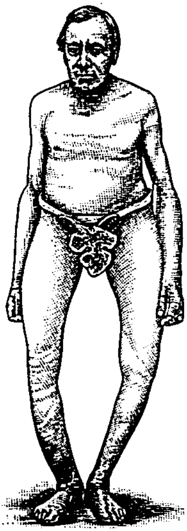

Fig. 11.

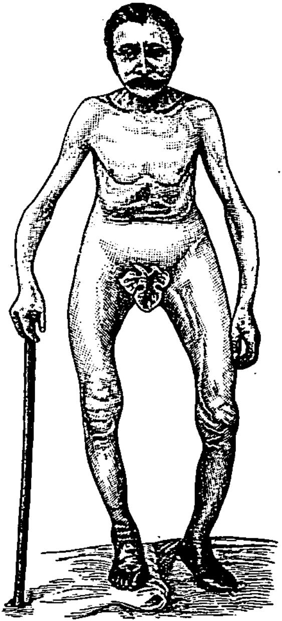

Fig. 10.—A. Robin's case. (Nouvelle Iconographie de la Salpêtrière, 1894.)
Fig. 11.—Gombault's case. Ibid.

So characteristic is the deformity in well-marked cases that it has seemed well for us to reproduce here Paget's original illustrations, with a few others from the literature. (See Figs. 6, 7, 8, 9, 10, and 11.) We would draw particular attention to the diminution in height, the curious deformity of the shape of the chest, the forward bowing of the head, the lozenge-shaped abdomen, the transverse sulcus running across the abdomen, the broadening of the pelvis, and the outward curvation of the lower extremities. To all these points attention has been drawn by previous writers. They make up a clinical picture which should not escape observation.

The relative frequency of malignant growths, which was first pointed out by Paget in his original communication, and which has been frequently mentioned by later writers as characteristic of the disease, would hardly seem borne out by a review of cases in the literature. Among them we find that 1 case (Clutton's) had a history of cancer in the family. Out of 66 cases, 3 had cancer. Including our own case, making a total of 67 cases, 5 had sarcoma; 2 cases had tumors of non-malignant nature. In 1 case (Lyon's) there was combined osteitis deformans and arthritis. From these facts it would seem that possibly too much stress has been laid on this association in the accounts found in the text-books. When we bear in mind that this disease is especially prone to affect those beyond middle age, it is not surprising that about 4.5 per cent. of the cases had cancer, although the high percentage of 7.5 per cent. of sarcoma must have some significance.

DIAGNOSIS. The chief difficulty in the matter of diagnosis is the determination as to whether a given case of deformity involving but one long bone is an incomplete case of osteitis deformans or whether it should be considered as a purely local affection. In some of the cases in the literature this question cannot be decided. The simultaneous involvement of more than one bone, particularly if the two bones be far removed from each other, and therefore not affected by a possible local cause, should make us suspicious of osteitis deformans, providing no manifest cause, such as syphilis or malignant growth, could be assigned. The differential diagnosis of this disease from others which apparently are somewhat closely allied to it, in being, so far as we can at present see, disorders of the growth of the skeleton, should not be a matter of difficulty. In mollities ossium, to which disease this seems certainly to bear some relation, there is not the same thickening of the bones as is seen in this disease, nor is the head involved. In fragilitas ossium we have deformity produced by the fractures which have taken place, but, except for the thickening around these fractures, there is no hypertrophy of bone. Acromegaly shows but few features in common with this disease, while it differs from it in the shape of the head, in the seat of the deformity and in the character of the latter. In osteitis deformans the cranium is triangular, with the base upward; in acromegaly the base is below, from the fact that in the former disease the calvarium is chiefly involved, while in the latter disease the lower jaw and orbital arches are involved far more than the calvarium. From leontiasis ossium this disease differs in the fact that the bones of the face are not affected, or only slightly, in osteitis deformans, while in hyperostosis cranii the long bones escape. To giantism this disease has not even superficial resemblance; in fact, the patient's stature is diminished owing to the bending of the long bones and to the almost constant dorsocervical kyphosis.

TREATMENT. Occasionally in the literature the statement is found that the pains accompanying the development of this disease seemed to be relieved by the use of iodide of potassium. In a far greater number of cases, however, the use of this drug has been productive of no relief, and, so far as we could find, there was no evidence of its producing any diminution of the size of the bones in cases which are evidently osteitis deformans and not simply osteitis due to syphilitic infection.

PATHOLOGY. The characteristic features of the morbid anatomy of osteitis deformans can be summarized as follows:

Skull. The distinction between the outer and inner table and the diploë is entirely lost. The entire thickness of the cranium is composed of finely porous bone substance, bounded on the inner and outer surfaces with a thin plate of harder bone. Microscopically the porous substance is seen to consist of a network of thin bony processes. The canals of the Haversian systems are enlarged and confluent, evidently the result of absorption. In the lacunæ, representing the normal medullary substance, are numerous giant cells, leucocytes, and fat cells, contained in a network of vascular connective tissue. Running through the affected areas, and seen as well in the healthy portions of the skull, when such are present, are bands of new-formed bony tissue. The osteoblasts of this new bone, while present in considerable numbers, are not as plenty as in healthy growing bone, and are irregularly arranged. The branching processes of the new canals are shorter than normal or are entirely lacking. The important characteristic of this attempt at regeneration is that the new substance remains uncalcified, and is in time reabsorbed. The border line between the new and old bone is sharply defined, indicating that the two processes are in no way connected. Most observers state that the regenerative process originates in the periosteum, or from the dura, while von Recklinghausen holds that it comes from the medulla. In our own sections the new tissue appears to come from the periosteum.

The result of the combination of these absorptive and regenerative processes is a total destruction of all symmetry in the internal structure of the skull or long bones.

As a rule, the presence of the uncalcified new bone substance renders the cranium softer than usual, but in some places areas of sclerotic bone tissue of ivory-like hardness are found.

In the long bones the normal relation of compact and cancellated structure is entirely destroyed. The outer walls of hard bone are represented by thin, irregular plates lying directly under the periosteum. In general, the histological picture here is the same as that of the skull, except that the medullary substance is more fatty, and that certain changes are observed that von Recklinghausen classes as retrogressive and progressive. The result of the former is the production of cysts filled with gelatinous substance. The progressive changes produce fibrous tumors or giant-celled sarcomata. The latter are sometimes found in the skull, as in the case here reported, but are rare in this position.

In the bodies of the vertebræ the histological changes are practically the same as in the skull.

A consideration of the morbid anatomy of the various bony structures affected in osteitis deformans readily explains the clinical features of the condition. The thickening of the cranium and of the long bones is due to the formation of a symmetrical new bone tissue. In the long bones and vertebræ the fact that this new tissue remains uncalcified and that the compact substance is being continually absorbed causes weakening, and the bending that results in the characteristic deformity of the disease. The tendency to fracture, seen in osteomalacia, is absent in osteitis deformans, since the presence of uncalcified new bone has a tendency to make the bones elastic and not easily broken.

Etiology. Up to the present time there is no thoroughly satisfactory explanation of the conditions necessary to produce the lesions of osteitis deformans. However, it may be of interest to briefly review the various theories that have been advanced. Von Recklinghausen holds that the process of absorption is a true osteomalacia associated with inflammatory processes, which commences synchronously with the destruction of the bony tissue, and leads to the transformation of the medullary substance into fibrous tissue. From this the processes of new bone grow out. The exciting cause, in his opinion, is mechanical. He bases his theory on the following facts:

1. That the cysts and sarcomata in the medulla, which he considers to be the first manifestation of the disease, occur at those points where the skeleton is most exposed to violence.

2. That the thickening occurs at the points of greatest strain—that is, in the shafts of the long bones and in the vertebræ.

3. That the skull is often affected, because it is exposed to extremes of heat and cold to a greater extent than is the rest of the body. These conditions act largely through their effect in producing contraction in the bloodvessels.

Paget thought that the whole process was inflammatory, associated with absorption, and explains the occurrence of the connective tissue in the medulla as a result of inflammatory hyperplasia.

Stilling thinks that the absorptive process is the result of a rarefying osteitis with its usual accompaniment, the formation of new bone which, however, remains uncalcified.

Lancereaux has advanced the theory that the new bone formation is an attempt at reinforcement of the long bones which have been weakened by absorption. This view is rendered untenable by the fact that the new tissue is formed not upon the concavity, but upon the convexity of the deformed shafts.

None of the last three authorities advance any theories as to the immediate cause of the disease. The theory that the exciting cause is mechanical renders it necessary to assume some predisposing condition, and up to the present time no satisfactory explanation has been advanced. It has been asserted by some that the process is a trophic one, associated with lesions in the nervous system. Stilling found a periependemal sclerosis in one of his cases. Von Recklinghausen describes masses of round cells around the bloodvessels of the cord in one instance. Gilles de la Tourette and Marinesco and Levi demonstrated sclerosis of the posterior and lateral columns and much degeneration of the bloodvessels of the cord. However, the absence of all clinical evidence of trouble in the nervous system and the results of many examinations that have shown the cord and bone nerves (von Recklinghausen) to be perfectly normal render it unlikely that the lesions referred to have anything to do with the origin of the condition.

DIAGNOSIS. The diagnosis between osteitis deformans and other affections of the skeleton of somewhat similar character is not difficult when based upon a study of the distribution of the lesions and their pathological histology. The disease most closely allied is osteomalacia; indeed, the two affections merge into one another. The main point of difference is that in osteitis deformans synchronously with the absorptive process a process of regeneration takes place. The callus that forms about the fractures that are so common in osteomalacia somewhat resembles the new bone of osteitis deformans, except that it is localized about the seat of a fracture. It often shows a lack of calcification. Moreover, fractures do not occur in osteitis deformans. The absorptive processes of both diseases are practically identical in their origin and appearance.

Leontiasis ossea affects the bones of the face, and the histological picture is that of osteo-sclerosis. The same may be said of hyperostosis cranii and gigantism.

Several writers upon the subject, especially Lancereaux and Richard, have made an effort to show that arthritis deformans and osteitis deformans were different manifestations of the same disease. They claim that both originate in some trophic disturbance, and in one the joints and in the other the shafts of the long bones and the skull are affected. Their evidence has been entirely clinical, and consists of several cases in which the two diseases appeared to be coexistent. No autopsy upon an undoubted case of osteitis deformans has borne out their theory.

Our conclusions from the study of this case and of the literature of osteitis deformans would be as follows:

A. From the Clinical Stand-point.

1. Beside our own case 66 true cases of osteitis deformans are found in the literature.

2. Osteitis deformans is a distinct disease of obscure etiology, but possibly allied to although not identical with osteomalacia, fragilitas ossium, and acromegaly.

3. The disease is one especially of later adult life, although its onset has been noted at as early a period as the twenty-first year. Of the 67 cases, 61 per cent. occurred in males, 35 per cent. in females. In a small number of cases trauma has seemed to play a part in the etiology. There is very little evidence of a family tendency to the disease, although there are a few examples in the literature.

4. The subjects of the disease bear a striking resemblance to each other in their general characteristics, the most noteworthy features of which are enlargement and forward projection of the head, dorso-cervical kyphosis, the prominence of the clavicles, the spreading of the base of the thorax, the diamond-shaped abdomen crossed by a deep sulcus, the relative increase in the width of the hips, and the outward and forward bowing of the legs.

5. The bones most frequently affected are those of the cranium, the tibiæ, and the femurs, in the two former of which the deformity was usually first noted. There is a curious preponderance of cases wherein the left side was either first or most involved, although at times it was noted that the enlargement was crossed so that the lower extremities of one side while the upper extremities of the other were involved to the greatest degree.

6. The association with malignant disease, while present in our case, would seem to be not quite so frequent as is usually stated.

B. From the Pathological Stand-point.

1. Osteitis deformans is a disease affecting the skull, vertebræ, and certain of the long bones. Its essential pathological characteristics are:

a. Absorption of the compact substance causing enlargement and confluence of the Haversian canals.

b. Formation of new bone which runs diffusely through the affected and the adjacent healthy portions. This new bone remains uncalcified, and is in turn reabsorbed.

c. The conversion of the medullary substance into a vascular connective tissue containing fat cells, giant cells, and leucocytes. In a small proportion of the reported cases cysts filled with gelatinous material and giant-celled sarcomata occur in the medulla.

d. As a consequence of these three processes, the ordinary relations of the compact substance and medulla are destroyed. The bones become exceedingly thickened and asymmetrical, but since the new bone tissue remains uncalcified its elasticity permits of great deformity of the long bones from the weight of the body, and fractures do not occur.

2. The whole picture of osteitis deformans from its pathological aspect is so very characteristic that it must be considered a distinct disease, and its pathological diagnosis is correspondingly easy.

3. The etiology of the condition is as obscure as when Paget first described it. Some predisposing tendency, probably trophic, must be assumed, and the exciting cause may be mechanical; in the skull, extremes of heat and cold, and in the vertebra and long bones the ordinary traumata to which these bones are exposed. Lesions of the nervous system are inconstant and rare, and are probably not a causal factor.

ASYMMETRY OF THE NASAL CAVITIES.[1]

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IN this community at least asymmetry of the nasal passages, to a greater or less degree, is so common that it might almost be considered the rule rather than the exception. It is found in those persons who practically never suffer from any disturbance of nasal function as well as in the patients in our clinics. It has been shown by Harrison Allen that asymmetry of the nasal passages may exist without deflection of the septum, but in the great majority of cases a deviation of the septum is the most prominent feature of the asymmetry. The etiology of these deviations, the forms which they may take, their frequency among different races, have been so often and so completely described that I need simply refer to papers by Ingals, Delavan, Bosworth, MacKenzie, Harrison Allen, and Roe, and to the works of Morrell McKenzie, Welcker, Ziem, Stoker, Rosenthal, Sendziac, McDonald, Collier, and especially Zuckerkandl.

In the literature of the subject of deviations of the septum and of asymmetry of the nasal fossæ are frequent references to changes in the shape of the turbinated bodies, commonly found in connection with

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[1] Read before the American Laryngological Association, 1901.